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CASE REPORT

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Difficulties in the diagnostics of renal tumor in a child with tuberous sclerosis – case report

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Summary

Background: Angiomyolipoma (AML) is the most common renal tumor in tuberous sclerosis (TS) patients. However, these patients also face an increased risk of malignant kidney neoplasms.

Case report: A 7-year-old boy with a diagnosis of TS was sent for abdominal CT due to a single isoechoic tumor in the lower pole of left kidney, which was revealed in US. CT exam confirmed the presence of a hiperdense tumor which exceeded margins of the left kidney. The tumor was homogenously enhanced after contrast administration. The absence of regions of negative attenuation in CT and the isoechogenisity in US suggested rather malignant lesion than AML. Partial nephrectomy was performed. Subsequent pathologic examination classified the tumor as a low-fat AML.

Conclusions: Radiological diagnosis of low-fat AML is difficult and requires a careful differentiation from other lesions.

Key words: tuberous sclerosis • angiomyolipoma • computed tomography

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Background

Tuberous sclerosis (TS) is a genetically determined disease that belongs to neuroectodermal dysplasia (phacomatosis) [1]. Patients are affected with organ changes (brain, skin, eyeballs heart, lungs, bones, kidneys), among which some show a tendency to malignant transformation [2, 3, 4]. The most common manifestations of tuberous sclerosis in kidneys are multiple bilateral changes in form of angiomyolipomas (AML) or cysts; clear cell carcinoma is also possible [1].

The aim of this work is to present a case of low-fat angiomyolipoma which was diagnosed during a prophylactic ultrasonography in a 7-year-old boy with TS.

Case report

A boy of 7 diagnosed with tuberous sclerosis was admitted to the Pediatric Surgery Clinic for further diagnostics and prospective treatment of renal tumor, which was found incidentally during a control USG examination of the abdomen. An ultrasonographic image of kidneys, acquired one year before, was normal. Check-up examination revealed a tumor of cortex echogenicity of 26x20x22 mm in the inferior pole of left kidney, which set off the outline of kidney and intussuscepted into its central echo.

Further diagnostics included Computed Tomography performed before and after intravenous injection of 2 ml/kg of contrast agent with the use of an automatic syringe. The acquisition after the injection was conducted in two phases: after 100 seconds and after 3 minutes. In the inferior pole of left kidney we found a well-circumscribed oval change, hyperdense in relation to renal parenchyma (61 HU; compare with renal parenchyma: 50 HU, muscles: 68 HU), which was homogenously enhanced up to 121 HU after the contrast medium administration (renal parenchyma: 160 HU, muscles: 84 HU). In the excretory phase the enhancement of the change

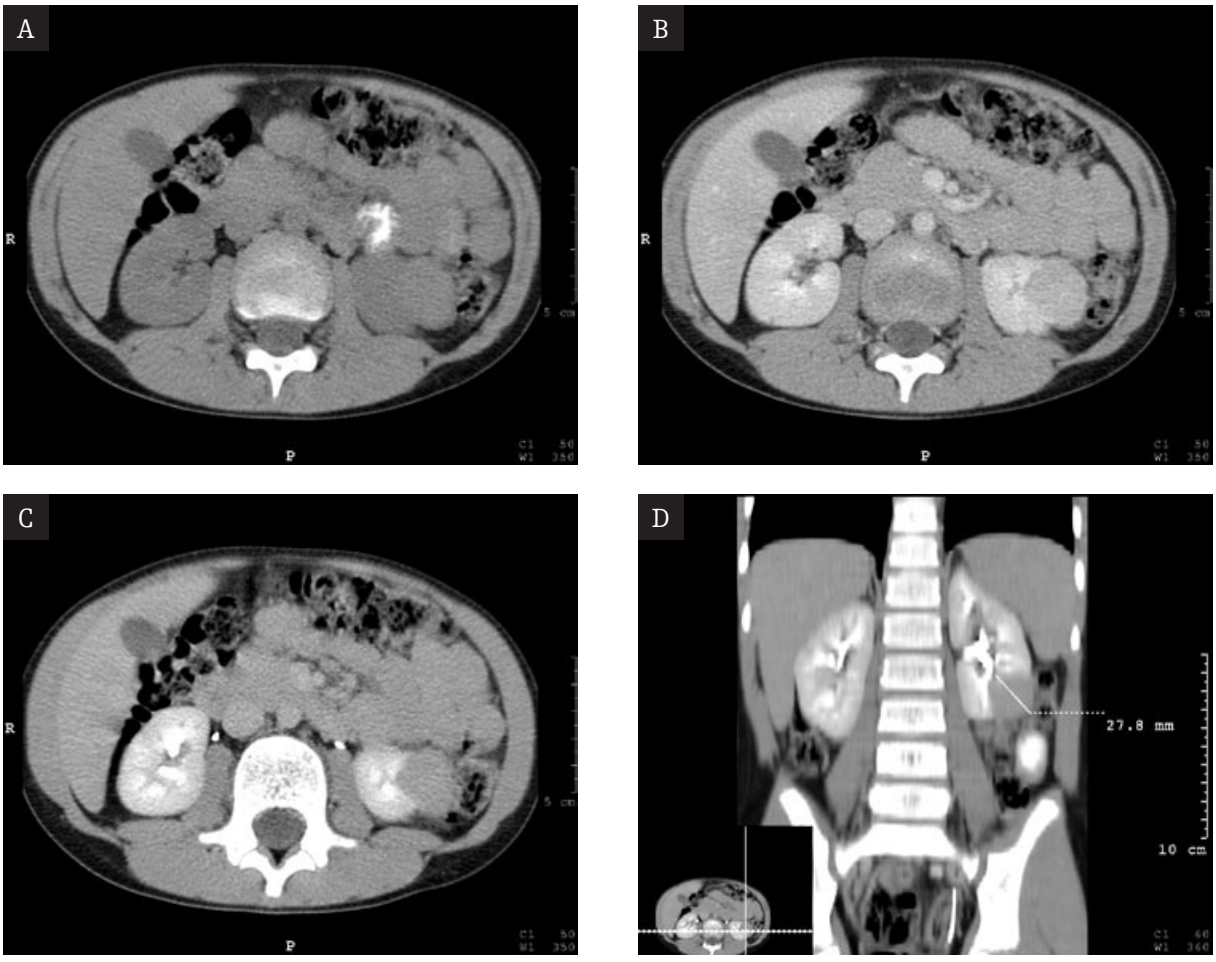

Figure 1. CT image of the low-fat angiomyolipoma: **A** – before contrast administration, **B** – parenchymal phase, **C** – excretory phase, **D** – MPR reconstruction in the frontal plane.

decreased to 94 HU [fig. 1]. The described lesion did not comprise any areas with the density of fat tissue. The tumor was tightly adjacent to pelvi-caliceal system and exceeded the margins of kidney. Apart from that, the image of kidneys in computed tomography was normal. In the conclusions of CT examination it was stated that the left kidney tumor can be equivalent to a malignant tumor. The child was qualified to a partial nephrectomy. Histopathological examination of the extracted change revealed that it was a benign mesenchymal tumor –a low-fat AML of 3 cm in diameter [fig. 2, 3].

Discussion

The most common pathological lesions in kidneys in course of tuberous sclerosis are multiple and bilateral changes in form of angiomyolipomas and cysts. Higher incidence of clear cell carcinoma compared to general population is also observed [1].

Renal AML occurs in 40–80% of all patients with tuberous sclerosis [1, 5]. Similar numbers are observed in children's population [6, 7, 8]. Moreover, such changes are usually multiple and bilateral [7], what in some cases makes a correct diagnosis easier to make. The average size of AML in children is about 2 cm [7]. In the described case we found a single tumor of 3 cm without related pathological changes in kidneys.

In microscopic examination the AML is built of fat tissue, disordered arrangement of vessels and smooth muscles [1, 5]. Owing to the presence of fat tissue, the presentations of these changes in imaging examinations are usually characteristic. In USG they would be hyperechogenic tumors, while in Computed Tomography – hypodense tumors with negative attenuation, which undergo contrast enhancement to different degrees, depending on the proportions of muscular and vascular tissues [7].

Diagnostic difficulties can be caused by angiomyolipomas with no or little fat tissue, described in literature as angioyoma or low-fat AML [9]. The incidence of this kind of tumor is estimated for 4,5% of cases [10]. Such changes are usually isoechogenic in USG examination, hyperdense in computed tomography with homogenous enhancement after the contrast injection [9–11]. However, these features are not pathognomonic [9]. The described low-fat AML appeared as a well-circumscribed isoechogenic tumor in ultrasonography. In CT, the lesion was homogenous, hyperdense in relation to renal parenchyma, and hypodense after the contrast injection. Lesions of negative attenuation which could be equivalent to the fat tissue, typical for AML, were not found.

Regarding the presented image, differentiation between low-fat AML and malignant renal tumors is crucial. In the

Figure 2. Macroscopic view of the tumor.

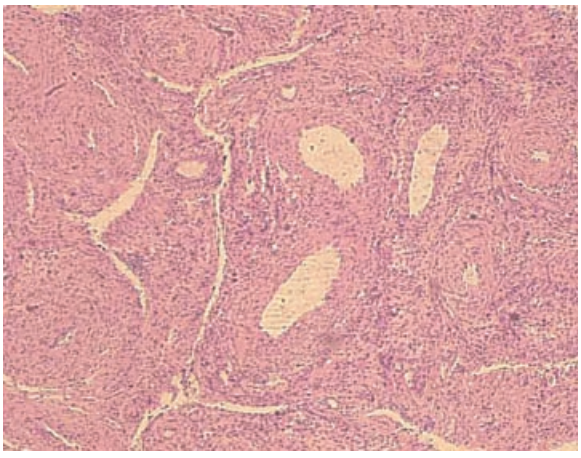

Figure 3. Microscopic image of the tumor (H+E staining).

described case the differentiating was focused on clear cell carcinoma in order to patient's age and diagnosed tuberous sclerosis. Both pathologies can have similar image in computed tomography, i.e. higher density compared to renal parenchyma before the contrast injection and homogenous enhancement, lower than normal parenchyma, after it [12]. According to several authors, reliable distinction of low-fat AML from clear cell carcinoma of kidney, based on imaging examinations, is not possible and requires histopathological verification [12, 9]. In such cases Patel et al. suggest to apply the cancer progression criterion to control examinations [12], although it requires cyclic CT scan. In our case, the tumor was found during annual ultrasonographic check-up, what did not allow an unequivocal assessment of its growth tempo in order to the lesion's isoechogenic structure.

Another malignant carcinoma that the AML ought to be differentiated from in the described case is a malignant angiomyolipoma. In rare cases, the changes defined as angiomyolipomas might exceed the renal capsule and occupy the regional lymph nodes [13]. The immunohistochemical examinations prove that it is a malignant form of AML [14]. In ultrasonography and computed tomography tumors of such type have image similar to AML and in this case the histopathological evaluation or growth observation is crucial as well.

In view of an ambiguous USG and CT image and inability to exclude a malignant process in the described patient, partial nephrectomy was carried out. *Tuberous Sclerosis Consensus Conference* recommends performing control USG examinations of kidneys every 1–3 years in children with tuberous sclerosis [15]. If a high dynamics of growth of the renal tumor is stated, its diameter exceeds 4 cm, CT examination of the abdomen ought to be performed. In case of any diagnostic doubts an open biopsy and alternatively – a partial nephrectomy should be carried out as well. Thin-needle biopsy is not recommended according to the risk of bleeding from the aneurysmally dilated vessels, which can disable renal functions and sometimes lead to nephrectomy [1].

Conclusions

Low-fat angiomyolipoma has an unspecific picture in imaging examinations and requires careful differentiation from clear cell carcinoma.

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